

CONGENITAL HYPERTROPHIC PYLORIC STENOSIS

A Historical, Anatomical, Pathological
and Clinical Study

by

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CONGENITAL HYPERTROPHIC PYLORIC STENOSIS.

Of the various terms used at one time or another to define this condition, Congenital Hypertrophic Pyloric Stenosis is the one most descriptive, and most nearly in accord with the theories of the majority of modern observers. At the outset we wish to make it clear that we are referring to a definite clinical entity in which there is invariably a demonstrable increase in thickness of the circular muscle layer.

HISTORICAL.

The disease is not a new one. Kellet recently calls attention to a case of pyloric stenosis in infancy, published almost a hundred years prior to any previously reported case. While it is by no means certain that this case represents a true example of congenital hypertrophic pyloric stenosis in infancy, it is suggested that its publication, which aroused a certain amount of interest, would have sufficed to have attracted attention to this condition had it been at all common at that time.

The case is reported by a German surgeon called Fabricius Hildanus in the year 1629. The child recovered on medical treatment, so no definite diagnosis could be made, but the history appears very typical. This /

This record appears to have attracted little attention as the next case reported is by Blair in 1717.

He describes a male infant, one month old, who was seized with violent vomiting and stoppage of urine and stool. Some time after, both these latter became more regular but the vomiting continued. He died at 5 months, very emaciated, weighing no more than five pounds. Post mortem revealed the ventriculus more like an intestine than a stomach, its length being 5 inches and its breadth but 1 inch. The pylorus and almost half the duodenum were cartilaginous and something inclined to ossification.

Christopher Weber reports a case in 1758, this time a female infant. Post mortem revealed a pylorus hard to touch, like cartilage, and contracted.

In 1771 George Armstrong reports the case of an infant, sex not stated, in which the post mortem examination revealed the condition as being due to spasm of the pylorus.

In 1778 Hezekiah Bardsley describes the clinical symptoms of necropsy findings in a child who died in the fifth year of life with symptoms dating from infancy. The pylorus was invested with a hard, compact substance, or schirrhosity, which so completely obstructed the passage into the duodenum as to admit with /

with the greatest difficulty the finest fluid. Whether this was the original disorder or only a consequence may, perhaps, be a question. He states, "In justice to myself I ought to maintain that I had pronounced a schirrhosity in that part for months before the child's death." We cannot fail to conclude from his description that this disease was exactly what we see today.

The literature seems to have suffered a gap of fifty years before any further literature on this subject was written. In 1841 Williamson of Leith and London reports a case under the title of "A case of Schirrhous of the stomach, probably congenital", which appeared to be a typical case in a male infant.

The following year Siemon-Dawosky recorded the first case in Germany and for the first time called attention to the projectile character of the vomiting. But these authors attributed the stenosis to hypertrophy of the sub-mucous layer.

The pathological anatomy as found in the adult was described by Landerer in 1879 and by Maier in 1885. Landerer was the first to use the term "congenital pyloric stenosis".

Their work was referred to by Hirschsprung in 1887, when he presented at Wiesbaden two specimens of the /

the condition in infancy, with observations on the clinical symptoms. The contributions of Hirschsprung in 1887 and 1888 actually initiated our modern interest in the disease in infancy.

The few cases reported in the decade following his article merely serve to confirm his observations.

In 1896 Finkelstein first called attention to the presence of a palpable tumour.

In the past twenty years the pathological and clinical pictures have been described by many observers; there is John Thomson's classical monograph in 1898 and others by Still, Holt, Sauer, Dorning, and Ibrahim. The valuation and widespread adoption, during this period, of a simple and effective operation has increased our knowledge of the pathological anatomy and has rendered diagnosis more exact. At the present time, the recognition of this condition is quite common, and it has been estimated that one in 200 male infants is affected.

It is interesting to trace the evolution of the operative treatment through the more complicated methods, such as jejunostomy, pylorotomy, division of the pyloric canal, gastro-enterostomy and pyloroplasty, down to the simple extra-mucosa partial pyloroplasty now generally known as the Fredet-Rammstedt operation. Some of the earlier operative procedures such as jejunostomy /

jejunostomy, as practised by Cordua, and resection of the pylorus as practised by Stiles, are now only of historical interest. Loreta's method of pyloric divulsion through a gastrostomy was never generally successful except in the hands of Burghard who had only one death in 46 private cases referred to him by Still. The first operation performed for this condition is credited to Cordua who, in 1893, performed a jejunostomy. In 1898 Maier and Stern each reported a case of pyloric stenosis in which gastro-enterostomy had been done. The first successful operation of gastro-enterostomy was performed by Lobker in 1898. This procedure apparently became the operation of choice and remained so for fifteen years. A number of cases were successfully treated by this method and, in the hands of a few exceptionally capable surgeons, the results were relatively satisfactory and the mortality not prohibitive. In the hands of most operators, however, the mortality remained high, with the result that surgical treatment was advised usually as a last resort and did not become generally popular until the more simple extra-mucous pyloroplasty was evolved. The first pyloroplasty after the method of Heinecke-Mikulicz involved a complete pylorotomy and was very difficult to perform on account of the thickness and hardness of the pyloric canal.

Nicoll of Glasgow then advised a V-shaped, sub-mucous pyloroplasty which he used in conjunction with Loreta's divulsion. In 1907 Fredet performed a successful sub-mucous pyloroplasty by means of a straight incision down to, but not through, the mucous membrane, subsequently transforming the longitudinal into a transverse wound by means of sutures.

Three years later, in 1910, Weber recommended the same procedure, calling it partial pyloroplasty. In 1912 Rammstedt called attention to the advantages of this operation, but omitted the transverse suture, and suggested leaving the pyloric wound gaping. This simplified procedure, now known as the Fredet-Rammstedt operation, has resulted in revolutionising the treatment of this condition. The results from this operation have been so favourable that, from being an operation of choice, it has now become almost the routine treatment for the condition.

Strauss advised an extra-mucous pyloroplasty which, in effect, was an elaboration of the Fredet-Rammstedt operation. In his procedure the mucous membrane is freed throughout its circumference and the exposed portion is covered with a flap derived from the thickened muscular layer. Experience shows, however, that the Fredet-Rammstedt operation has proved so unanimously satisfactory that we have seen no reason to employ any variation.

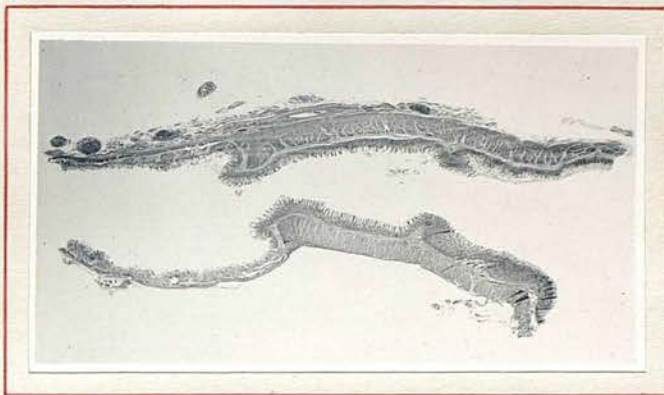
ANATOMY.

The normal pyloric canal in the infant presents itself as a cylinder approximately 1.5 cm. in length. In the infant it is generally found to be contracted and its lumen obliterated by closely packed longitudinal folds of mucous membrane. In the adult it is more common to find the canal partly expanded, so that its demarcation from the pyloric vestibule is less marked. The extremity of the pyloric canal protrudes into the commencement of the duodenum as a smooth rounded knob, surrounded by a shallow fornix, and having in the centre a small puckered orifice, the pyloric opening. The appearance is that of a miniature portio vaginalis of the cervix uteri, a resemblance more marked in the infant than in the adult. The pyloric canal is provided with a powerful sphincteric apparatus. Both the circular and longitudinal muscular fibres are present in a greater mass than in any other portion of the stomach. The circular fibres are disposed in the form of a thick muscular cylinder which surrounds the entire length of the canal. At the duodeno-pyloric constriction the margin of this cylinder becomes increased in thickness, forming thereby a strong muscular ring which encircles the pyloric orifice and constitutes the pyloric-sphincteric ring. There /

There is no sudden or abrupt transition between the muscular substance of the pyloric canal and that of the sphincteric ring. The fasciculi of the latter are, however, quite cut off from the muscular coat of the duodenum by a distinct connective tissue septum. During life the opening of the pyloric canal into the duodenum is closed, except during digestion, when it opens at irregular intervals to allow the contents of the stomach to pass into the duodenum. Clinical evidence seems to suggest that, under certain circumstances, the entire muscular cylinder is employed as a sphincter, and thereby the entire length of the pyloric canal acts against the entrance of material from the stomach. Longitudinal muscle fibres radiate from the oesophagus in all directions over the stomach, ending, however, before they reach the pyloric portion. A new set of longitudinal fibres takes origin in the body of the stomach, and these become more numerous as they continue over the pyloric canal, forming a distinct longitudinal coat. Comparatively few of these fibres pass over to become continuous with the corresponding fibres of the muscular coat of the duodenum. The deeper longitudinal fibres leave the surface and penetrate the substance of the pyloric-sphincteric ring. Thus is formed an effective apparatus antagonistic /

antagonistic to the pyloric-sphincteric ring, by means of which the pyloric orifice may be dilated when the nervous mechanism concerned in inhibition brings about a relaxation of the sphincter. There is then both a constrictor and a dilator of the pylorus.

Fig. A. Section of the normal pylorus of a child of 9 weeks.



Note. 1. The normal circular muscle of the pyloric canal.
 2. The thin longitudinal muscular coat.
 3. The normal appearance of the mucous membrane in the pyloric antrum and the appearance of the sphincter which forms a slight projection into the duodenum and /

and results in the appearance of a miniature portio vaginalis of the cervix uteri.

Fig. B. Magnification 10 times.



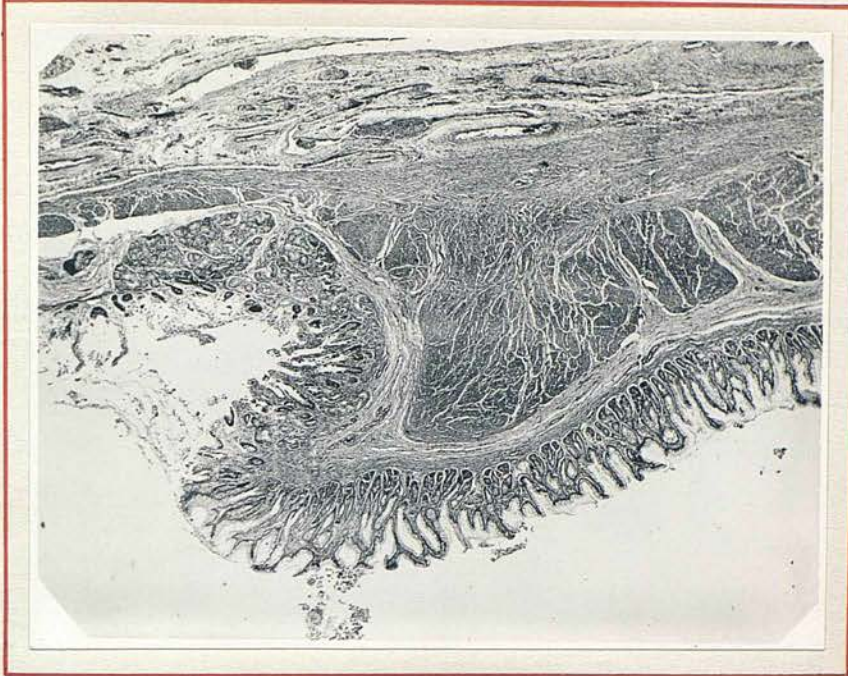
- Note.
1. Termination of the longitudinal muscle fibres in the pyloric sphincter.
 2. The fasciculi of the pyloric sphincter are cut off from the muscular coat of the duodenum by a connective tissue septum.

Fig. C. Magnification 15 times.



Note. Though no definite line of demarcation can be made out between the muscular substance of the pyloric canal and that of the sphincteric ring, there is, however, a very definite difference in the appearance of the muscle fibres of the pyloric canal. They appear to be cut, on cross section, and stain slightly deeper.

Fig. D. Magnification 25 times.



Note. This shows a further magnification and there seems to be slight evidence of a line of demarcation forming between the sphincter and the circular muscle fibre. It is to be observed that the pyloric sphincter is covered by normal gastric mucosa and only in the angle formed by the projection of the sphincter into the duodenum are Brunner's glands seen, the sphincter itself being covered by gastric mucosa.

Crymble and Walmsley of Belfast report a case, in the British Journal of Surgery, 1933, of pyloric hypertrophy in the adult. They believe this case to be a persistence of infantile hypertrophy, and suggest re-examination of the pathological anatomy of infantile hypertrophy of the pylorus considering that hypertrophy does not affect the pyloric sphincter, which now remains under-developed, but is only of the muscle wall of the whole length of the pyloric canal. They state further communications will be given on this matter but as yet nothing has appeared in print. The pathological sections which they exhibit have stimulated us to investigate this condition in the infant.

Following on Crymble and Walmsley's publication, Twining of Manchester reports three cases of hypertrophic stenosis of the pylorus in adults and believes he is able to demonstrate radiologically the presence of a pyloric sphincter and prepyloric hypertrophy.

We /

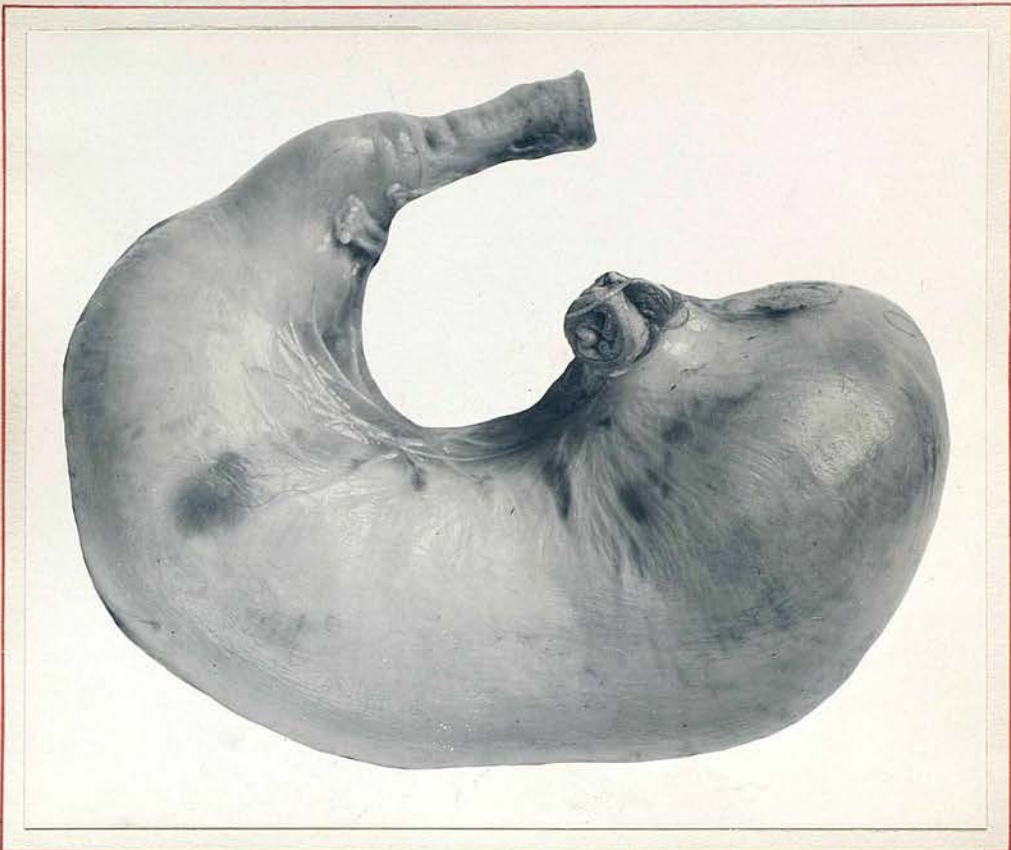
We have been fortunate enough to obtain the stomach of a child who died at the age of three months, having suffered from vomiting and constipation since the fifth week after birth. The child weighed five pounds at birth, appeared to put on weight normally while on the breast, but started vomiting at the fifth week. This vomiting was associated with constipation, and the family doctor took the child off the breast and gave special artificial feeding. The child, however, continued to vomit, though at times the constipation appeared a little less severe. It was admitted to the Children's Hospital, Edinburgh, at the age of three months. The child on admission was extremely collapsed and suffering from severe malnutrition. Four-hourly intravenous salines were given and lactic acid and skim milk feeds. However, little response was effected, owing to the serious condition of the child on admission, and it died 48 hours later. At post mortem a typical case of congenital hypertrophic stenosis of the pylorus was found. Miss MacGregor, Pathologist to the Children's Hospital, very kindly gave us the stomach and we have made sections which will be discussed in detail, hoping to prove that in the infantile pyloric hypertrophy, as well as in those cases discovered in the adult and attributed to persistence of this condition, there is no hypertrophy of the pyloric sphincter.

PATHOLOGY.

The appearance of the pylorus in a well-defined case is so characteristic and uniform that there is a striking similarity in description by various authors. The pylorus has the feel of a solid cylinder about 2.5 cm. in length. The size of this tumour varies from that of the distal phalanx of the little finger to that of the thumb. During muscular contraction the consistency of the mass is of almost cartilagenous hardness and the colour is white. During relaxation these characteristics are not so marked, but at all times the tumour is firm and paler in colour than the stomach and duodenum. The lumen of the pyloric canal is gradually narrowed but will always admit the point of a probe. On cross section, the pylorus is found to be much thickened, and this thickening is caused by a hyperplasia of the circular muscular layer. The sub-mucosa is often oedematous. The other layers are unchanged. The increase in thickness involves chiefly the circular muscular layer of the pyloric canal, and we will try to show that the sphincteric ring is slightly, if at all, involved in the hypertrophy. The over-lying peritoneum is smooth and glistening, and the tumour is freely movable. As a secondary change the mucous membrane /

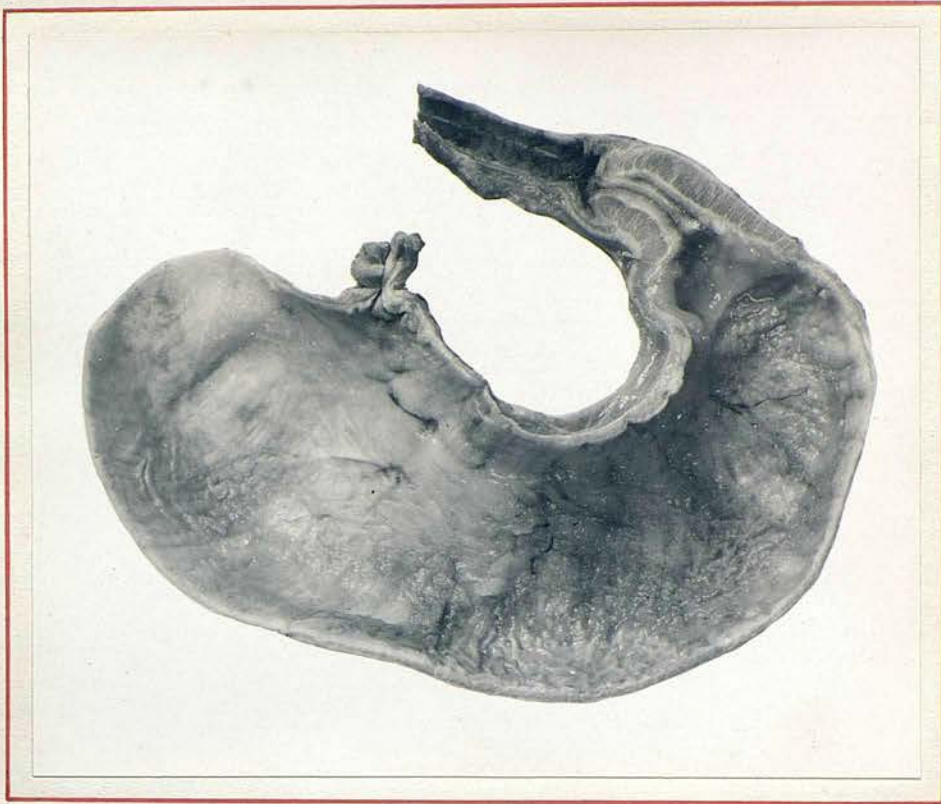
membrane of the stomach shows some degree of gastritis with hypertrophy of the walls and dilatation of the cavity of the stomach depending upon the degree of obstruction.

Fig. I. This Figure shows the stomach of a child three months old who suffered from congenital pyloric stenosis.



- Note.
1. The markedly dilated stomach.
 2. The ovoid, hard, pyloric tumour.
 3. The small, contracted duodenum.
 4. Interlacing of longitudinal circular muscle fibres over the cardiac end of the stomach.
 5. The smooth overlying peritoneum.

Fig. II. Longitudinal section of previous stomach.



- Note. 1. Dilated stomach cavity.
2. Hypertrophy of stomach walls.
3. Oedematous sub-mucosa with excessive mucus
in pyloric antrum.
4. Much thickened pylorus as a result of hyper-
plasia of circular muscle fibres.
5. Thinned out longitudinal muscle fibres over
the tumour.
6. Narrowed pyloric lumen.

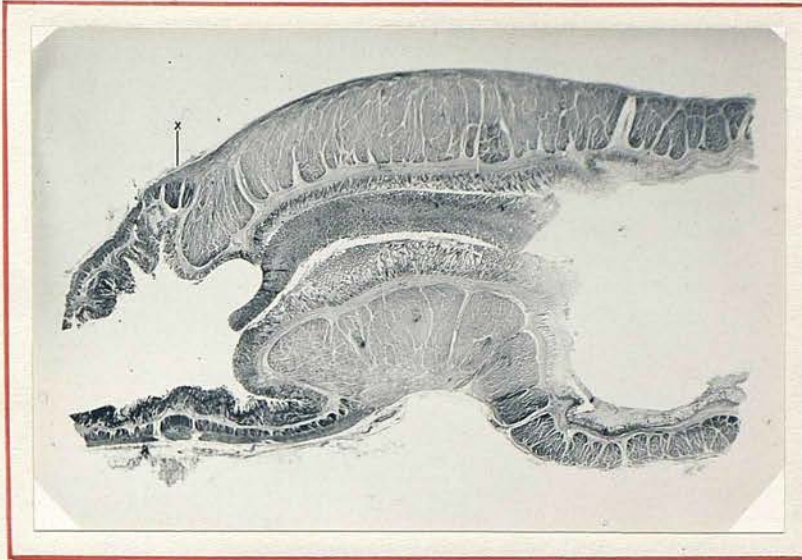
PATHOLOGICAL SPECIMEN.

Longitudinal Section. Longitudinal section, from the pyloric end of the stomach parallel with, and close to, the greater curvature, shows great hypertrophy of the circular muscle which formed the tumour, the sudden commencement of the hypertrophy, and that it exists in the whole length of, and is confined to, the pyloric canal. It shows, too, that the longitudinal muscle is thick over the pyloric antrum, very much thinner over the tumour, in fact, rather less than normal. It shows, also, in the pyloric antrum, the mucous membrane is folded and the submucous layer is increased in amount, loose and oedematous, and that in this region there is a uniform, thickened, circular muscle coat, and that there is a distinct, thickened, longitudinal muscle coat. The mucous membrane over the tumour is practically unfolded and it is obvious that it is much more closely and firmly bound to the muscle. The duodenal mucous membrane covers the whole of the duodenal face of the tumour.

The hypertrophied areas consist of bundles of normal visceral muscle which are looser in their arrangement, and embedded in a greater amount of fibrous tissue than is usual in smooth muscle. The fibrous tissue is continuous with that of the submucus layer. In the internal parts of the mass the bundles /

bundles are almost entirely circular but more superficial, they are oblique or vertical, and here they freely interlace on one another. These bundles are continuous with, and appear to be extensions of, a more or less normal layer of circular muscle which underlies the thin layer of longitudinal fibres on the surface. In the position where the normal sphincter would be, however, and over an extent that would correspond with the normal sphincter, the normal circular fibres are thickened, as they would be in a normal, though small, pyloric sphincter, and over this sphincteric thickening the superficial longitudinal layer almost disappears as such, as it does over the normal sphincter. It is further to be noted that this pyloric sphincter makes no contribution to the hypertrophied mass and is, in fact, separated from the duodenal end of the mass by a layer which is seen to be continuous with the sub-mucous and muscularis mucosa.

Fig. III. Micro-photograph of pyloric antrum, pylorus and duodenum of previous specimen. Magnification: Multiply by 3.



In the above section the following points are worthy of note:

1. Great hypertrophy of the circular pyloric muscle which forms the tumour.
2. Thinned longitudinal muscle layer over tumour.
3. Ending of longitudinal muscle layer in the pyloric sphincter marked with a X.
4. Pyloric sphincter separated from the duodenum by a layer which is continuous with the sub-mucus and muscularis mucosae of the duodenum and separated from the circular /

circular pyloric mass by a few strands of longitudinal muscle running to be continuous with the muscular mucosa covering the projecting hypertrophied muscle.

5. Duodenal mucosa with Brunner's glands covering duodenal surface.
6. Area known as "Zone of Adhesion" where muscle mucosa and serosa are bound into an almost inseparable mass.
7. Normal duodenal muscle with absence of any longitudinal fibres passing from stomach to duodenum.

Fig. IV. Magnification of the previous photograph taken at the fornix caused by the protrusion of the pylorus into the duodenum. Magnification 10 times.



- Note. 1. The staining fibres of the pyloric sphincter, which appear to be cut off from the hypertrophied mass of circular muscle fibres by a few strands of longitudinal muscle fibres which run to be continuous with the muscularis mucosa.
2. The end of the longitudinal muscle appears to terminate in the pyloric sphincter /

sphincter.

3. The longitudinal muscle fibres terminate here abruptly and few pass over to the duodenum.
4. The sphincter pylorus is separated from the duodenal musculature by a few strands of muscularis mucosa.

Fig. V. Further Magnification of above section - (multiply by 15). This section shows more clearly than ever the distinct pyloric sphincter. It is seen to be covered by duodenal mucosa and Brunner's glands.

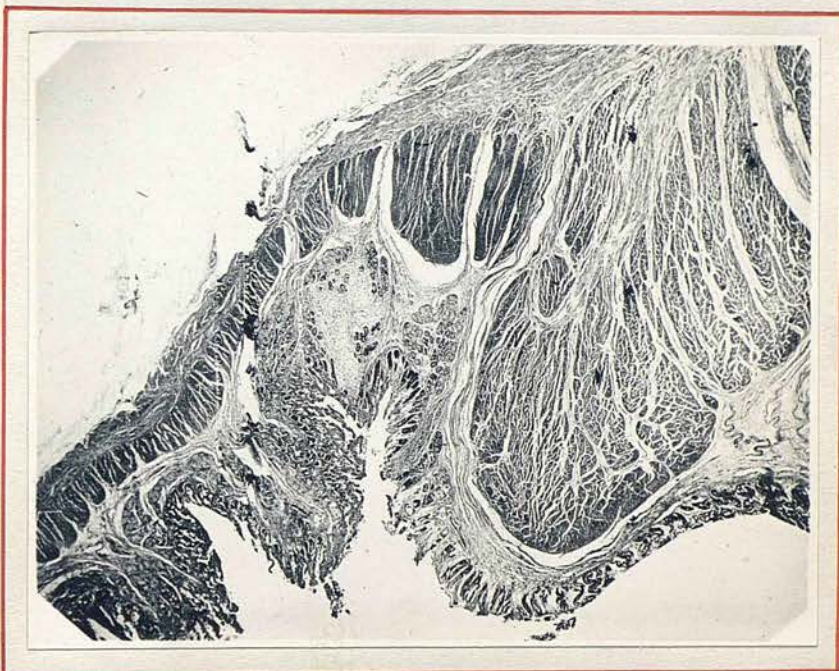


Fig. VI. Further Magnification - (multiply by 25). Here the pyloric sphincter appears to stand out most distinctly. Its muscular fibres do not appear to be of the same texture as those of the hypertrophied circular muscle. They are more closely bound together and stain differently in comparison to the looser hypertrophied bundles.

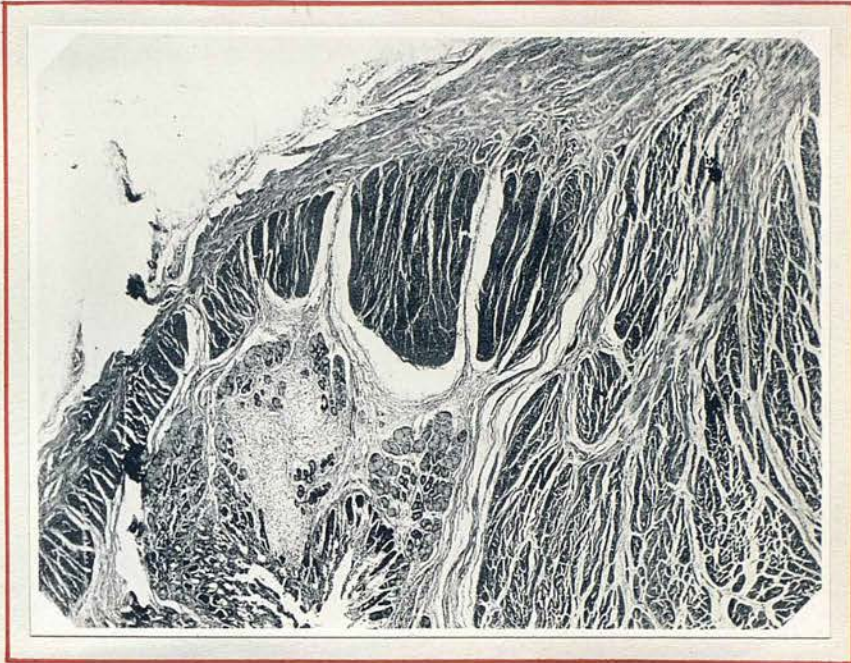


Fig. VII. (After Thomson & Findlay). Shows a cross section of a normal and a hypertrophied pyloric canal.

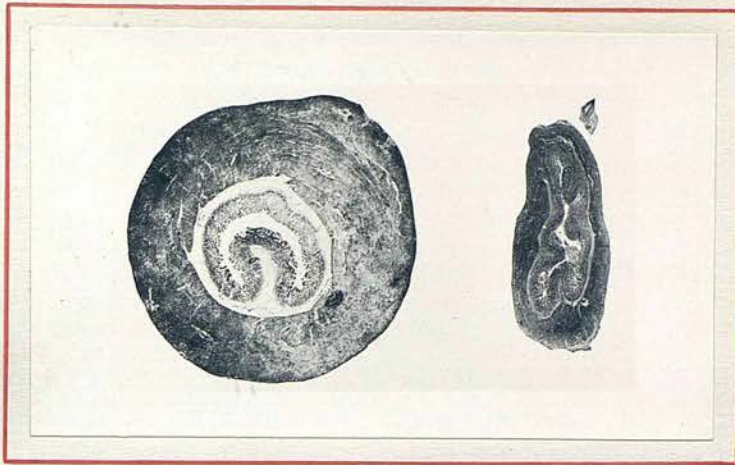
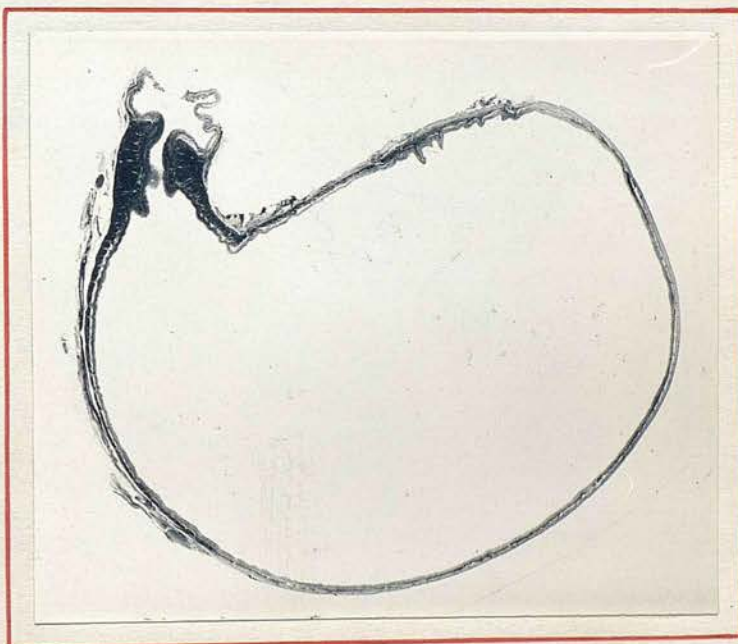


Fig. VIII. Whole section of a stomach showing evidence of pyloric stenosis. It is from a rather old specimen and did not show up essential details very well. Unfortunately the whole section of this case was cut obliquely, failing to establish a pyloric lumen and so useless for reporting purposes.

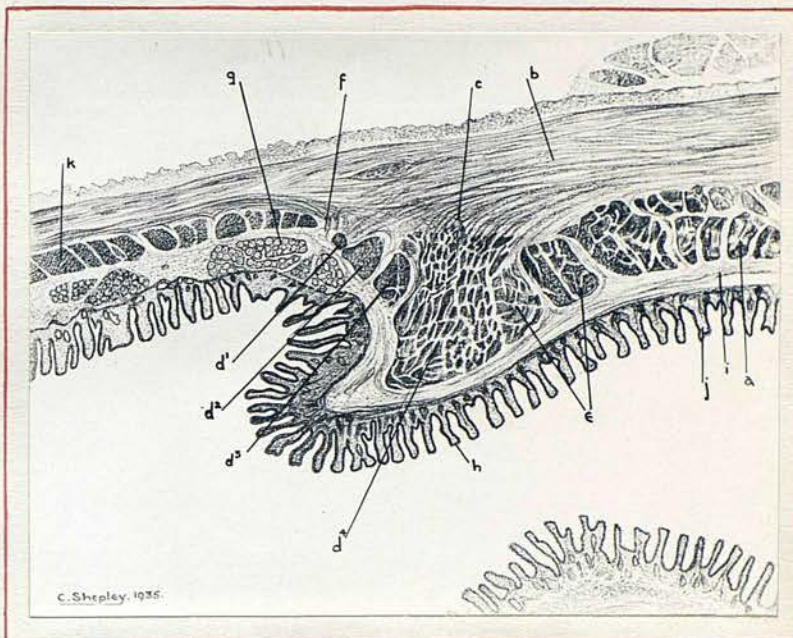


In order to illustrate the points which we consider of importance in these sections, we had illustrations made of the actual sections, and the main areas labelled.

Drawings have been made from microphotographs.

If we consider the normal illustration first we see:

Fig. E.

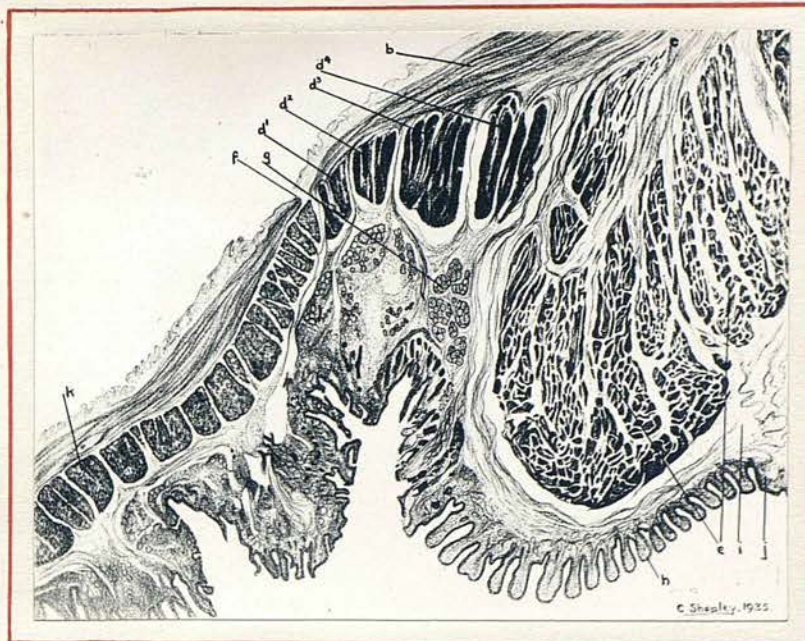


- a. Normal circular muscle of pyloric canal.
- b. Longitudinal muscle coat.
- c. Longitudinal muscle fibres are seen to terminate in the pyloric sphincter.
- d. Is composed of d_1 , d_2 , d_3 , and d_4 , all these portions go to make up the normal pyloric sphincter.

e /

- e. Here, we believe, a difference in the structure of the pyloric sphincter and the circular muscle fibres can just be made out. There is no clear line of demarcation, as would be expected, but it appears as if there is a difference in d_4 , the major portion of the pyloric sphincter, and the portion of tissue on its right marked "e."
- f. Here, on the other hand, is a definitely clear demarcation line between the pyloric sphincter and the circular duodenal muscle coat.
- g. Duodenal glands or Brunner's glands.
- h. and j. Normal gastric mucosa occupying the pyloric canal and covering the sphincter down to the fornix.
- i. Muscularis mucosa.
- k. Circular muscle coat of duodenum.

If we now consider the illustration of the same area of the pyloric canal in a child, practically the same age, who died of congenital pyloric stenosis, we observe:

Fig. F.

b. The longitudinal muscle fibres running to terminate in the pyloric sphincter d.

d. Again is composed of d_1 , d_2 , d_3 , and d_4 . It will be seen that the sphincter stains darker than the hypertrophied circular muscle "e" and is about the same size as the normal sphincter.

e. It is our opinion that "e", as seen in the normal illustration, has become markedly hypertrophied, has grown over the sphincter, and compressed the sphincter higher up into the fornix. The fornix has become larger from the projection of this increased mass of circular muscle tissue of the prepyloric canal into the duodenum.

f. Line of demarcation between pyloric sphincter and duodenal /

duodenal musculature.

- h. and j. Thinned out gastric mucosa covering hypertrophied circular muscle.
- i. Muscularis mucosa.
- k. Circular duodenal muscular coat.

Crymble and Walmsley ask for a further investigation of the pathology of congenital pyloric stenosis, feeling that if in their case, which was a persistence of the congenital hypertrophy in early adult life, a definite pyloric sphincter was demonstrated, that this point should be investigated in the infant.

We feel that in this small number of cases investigated we have been fortunate in being able to show that the hypertrophy is confined to the prepyloric canal and not the pyloric sphincter.

This latter structure is slightly, if at all, affected by the general hypertrophy.

SURGICAL ANATOMY.

This has been admirably discussed by Dennis Browne and it is difficult to do better than to use his classification. The structures encountered in freeing of the pylorus fall into five main groups.

(1) Serous Coat. This has no particular features to distinguish it from the visceral peritoneum covering the alimentary tract elsewhere. Features which are important, however, are that it is extremely adherent to the muscle beneath, and it is tough and inextensible.

(2) Blood Vessels. These run from two sources, the right gastric and gastro-epiploic vessels, and in two planes, a superficial and a deep. The superficial ones show conspicuously on the surface under the serosa, but in the region of the operation they fail to make any visible anastomosis at the midpoint from their origins. The hiatus between them forms the well-known Bloodless Line which follows the axis of the viscus and so is curved upwards at its left extremity. The deeper vessels mostly ramify in the mucosa, but there are two which are almost invariably seen in the muscle during the operation. The most important of these is the artery with its accompanying vein which is met running across the duodenal end of the incision, just at the apex of the fornix of the mucosa /

mucosa to which it forms a very useful indication. The other vessel appears in the deeper layers of the muscle, towards the other end of the wound, and occasionally causes trouble from bleeding.

(3) Muscular Coat. The only pathological change in the disease is an overgrowth of the muscular fibres, especially at the pylorus, and this results in blockage which is mechanical as well as functional. This swelling of muscle takes place inside an adherent and unstretchable covering of peritoneum, and consequently can go in two directions only - inwards into the lumen and lengthways, mainly into the duodenum. The result of the swelling inwards is that the pylorus, instead of being, when relaxed, the normal and patent tube, remains a hard, solid mass. The swelling downwards produces protrusion of the pyloric orifice into the duodenum. Another mechanical effect of the confinement and hypertrophy of the muscle is a decrease in its vascularity from the compression of the vessels supplying its fibres, and, finally, the muscle is changed in its consistency, becoming more hard and cartilagenous.

(4) Areolar or Sub-mucus Coat. This inconspicuous layer makes the operation possible, and, to understand its relation, it is necessary to study its functions. Here, as elsewhere in the body, the areolar /

areolar tissue exists to form an action that will allow two structures to move freely on one another, and, as usual, one of these structures is muscle. The other is the mucosa, and the need for independent movement arises because it, being non-contraction, must meet the contractions of the surrounding muscle by wrinkling into folds inside it. As might be expected, the amount in looseness of the areolar tissue, and the depths of the folds, are greatest at the points where variations in the diameter of the tube are greatest, and least where they are least. Thus the areolar layer is well-marked in the pyloric antrum, becomes still wider and looser up the pyloric canal, and decreases markedly over the projection of the sphincter, ceasing entirely at the apex of the fornix, and then reappearing in the duodenum.

Fig. IX. (After Dennis Brown).

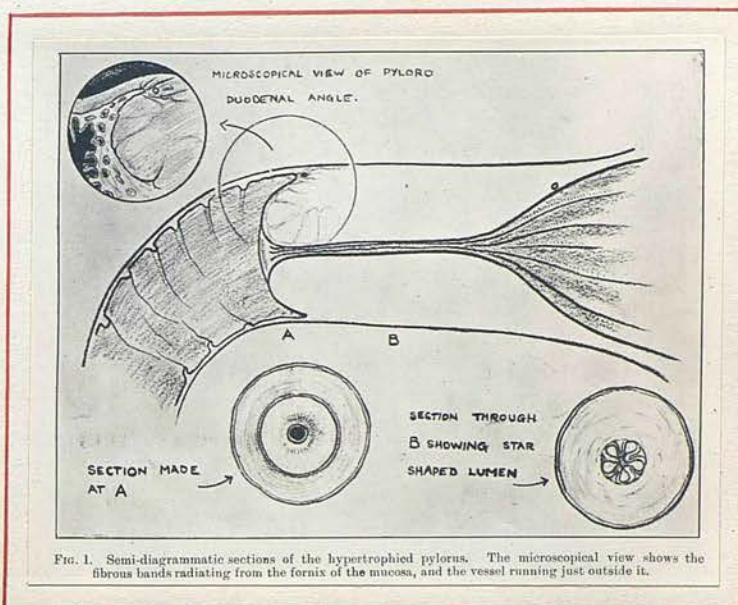


FIG. 1. Semi-diagrammatic sections of the hypertrophied pylorus. The microscopical view shows the fibrous bands radiating from the fornix of the mucosa, and the vessel running just outside it.

(5) The Mucosa. This is the most important of all layers and the most difficult to understand and describe. In the pyloric antrum it is of considerable thickness and is wrinkled into longitudinal folds which increase in depth as the pyloric canal is reached and so give the well-known star-shape of the potential cavity which is seen here on cross section. These deep folds run through the canal till the actual opening in the duodenum is reached, when they suddenly cease, just as a bore of muscular tube starts to widen. Consequently, at the apex of the pyloric protrusion into the duodenum, although the actual diameter of the mucous tube is far greater than that at the pyloric canal, the potential diameter is far less. In other words, if the entire mucosa in this region be freed of muscle and dilated to its full extent it will be found that, at the pyloric orifice, there is a very marked choke in its bore, like that at the muzzle of some shotgun barrels. At the end of this zone of constriction, which only lasts for about an eighth of an inch, the mucous membrane suddenly expands into the full diameter of the duodenum, which is approximately twice that of the choke, and the same as that of the potential diameter of the pyloric portion. It rises backwards, smoothly applied to the protruding nozzle of /

of muscle, to form the fornix already mentioned, and then forward again as the lining of the duodenum. On consideration it will be seen that the apex of this fornix is subject to a considerable force tending to tear it downwards into the duodenum whenever the stomach contracts against the stopper-like mass of muscle in the pylorus. As might be expected, this force is counteracted by the development of fibrous tissue, as in the similar case of the tonsil. Fibres run just as Lane's doctrine of the organisation of lines of force would lead one to expect, outwards and downwards from the angle of reflection of the mucosa into the muscle. In consequence a "zone of adhesion" is formed at this point in which mucosa, muscle and serosa are bound into one almost inseparable mass, but, immediately beyond it, the mucosa once again becomes rugose and lies loosely attached to the duodenal muscle. In this area the mucous membrane is a very vascular and vital structure which can be stripped of all protection over a large area without any risk of sloughing. It is also, like all sheets of tissue, of surprising tensile strength as long as it remains uninjured, but once it has been punctured the tear will extend with the greatest readiness.

Fig. X /

Fig. X. (After Dennis Browne).



ETIOLOGY.

The etiology has given rise to endless discussion and remains obscure. Hirschsprung's theory was that there was a primary developmental hyperplasia of the circular muscle fibres of the pyloric ring and canal, and that the reduction of the pyloric lumen by the increased thickness of the muscle layer was the cause of the obstruction. In this connection it is of interest to note that Wernstedt attempts to explain these pyloric hypertrophies of infants on the basis of reversion to a type of sphincter found in the Edentata. Most observers now agree that in every /

every clinical case there is a certain amount of spasm. The hypertrophy alone apparently does not produce sufficient narrowing to cause symptoms in infants. The relation of spasm to hypertrophy, and of each to symptoms, has been the subject of a mass of, so far, futile conjecture, and of much unresolved controversy. The adherents of the spasm theory believe that the primary disturbance is not an anatomic anomaly but a spasm of the muscle fibres producing a fundamental narrowing of the lumen, with more or less subsequent hypertrophy of the muscle layer. There is no evidence that spasm can cause hypertrophy.

Spasm of the pylorus does occur at all ages, but it does not produce the condition of hypertrophy which is, in general, peculiar to the first few weeks of life. John Thomson, of Edinburgh, was of the opinion that there is some congenital lack of stability in the harmony of gastric and pyloric relaxation and contraction, so that the child starts life with a machine which is certain to get out of order. John Fraser supports this view. Finally, there is the theory which assumes the hypertrophy to be primary and the spasm secondary. In our opinion there is a primary, developmental hyperplasia of the circular muscle fibres of the pyloric canal. The belief /

belief that this hyperplasia is congenital is supported by the fact that the condition has been noted in a seven months' foetus and, in several instances, within a few days or hours of birth (Dent, Greenbaum, MacHaffie). Associated with this hyperplasia is a secondary, increased, functional activity and the combination results in pyloric obstruction. In any event, we are of the opinion that hypertrophy precedes spasm in all cases. Yet not until spasm has been added do symptoms appear.

Whether this is a true spasm, or merely a lack of co-ordination between the nerves controlling the movements of the stomach and the opening and closing of the pylorus, is not known.

Whichever of these latter conditions obtains is the one which is responsible for the production of symptoms. That this element of spasm is the cause of symptoms is evidenced by the fact that symptoms are rarely noted during the first two weeks of life, and that, after the condition has manifested itself, periods of remission may be observed during which the baby does not vomit. The severity of the symptoms is dependent on the degree of spasm and the amount of obstruction caused by the hypertrophy of the circular muscle layer. There is no relationship between the severity of the clinical symptoms and the actual size/

size of the tumour.

Pyloric Inco-ordination. An interesting article reviews the opinions of Pritchard and Levi who consider the terminology of this disease is wrong and it should be referred to as an inco-ordination. Pritchard, who is credited with being responsible for the suggestion that these children suffer from an abnormality of the gastro-duodenal reflex, states that the actual phase of the pyloric reflex is that of relaxation and not of closure of the sphincter.

PYLORIC THEORIES.

The normal, or inactive phase of all sphincters, such as the pyloric, the caecal, or orectal, is to be in a mild condition of tonicity. When the effective reflex comes into play there is relaxation.

It is a great mistake to speak of stenosis at all. Of all the innumerable patients examined post mortem we have never yet seen a case of true stenosis of the sphincter opening, even though the hypertrophy on the gastric side may have been enormous.

A sphincter is either open or shut - it is never in the halfway position.

Failure of the stomach to empty itself is not due to stenosis. It is due to failure of the sphincter /

sphincter to relax.

Instances in which there is obstruction immediately after birth are, in our experience, remarkably rare, although the obstruction soon becomes manifest owing to failure of acquisition of the normal reflex.

Many infants at birth do suffer from abnormality of their reflexes.

Some reflexes are slow to appear. For instance, the oesophageal reflex may be under-developed. Difficulty is then experienced in persuading the child to swallow.

Assuming a late development of the reflex which opens the pylorus, it does not require a very wide stretch of imagination to visualise an imperfectly balanced mechanism which would send the pylorus into spasm and which would subsequently lead to hypertrophy of the muscle. Development of muscle occurs very rapidly in infants.

This assumption explains how food is enabled to trickle through the pylorus during the first few weeks of life. How it is held back in the stomach only when the muscle has become spastic and subsequently hypertrophic. This theory also brings into line two separate conditions, which have been described, pyloric spasm and pyloric stenosis.

Pyloric /

Pyloric spasm in infants appears to be but an early manifestation of so-called pyloric stenosis. Some infants with pyloric spasm develop their normal gastric reflex and get well spontaneously. In others the reflex is late in appearing and the muscle hypertrophies.

They thus become examples of pyloric stenosis. Instances have occurred in premature children. The foetus may drink liquor amnii and there is no logical reason why, in some rare instances, the pylorus should not hypertrophy before birth, being stimulated to do so by the abnormal peristalsis set up as a result of the ingested fluid. Pyloric spasm and pyloric stenosis may then be results of this underdevelopment of the gastric duodenal reflex, and both lesions should be classified together as examples of pyloric inco-ordination.

SYMPTOMS.

The clinical picture is a striking one and, in the majority of cases seen, is remarkably uniform. We have in our experience found no evidence of racial predisposition, the condition being observed in Europeans, non-Europeans, Asiatics. None but Europeans, however, have consented to any form of surgical treatment.

The /

The clinical history is very typical. An infant of three to four weeks old, who has hitherto shown no tendency to vomit, or perhaps has posseted a mouthful or two, begins to vomit occasionally. The vomiting is not necessarily very frequent. Perhaps only twice in twenty-four hours. The vomit is large and consists of more than the last feed. Moreover, it comes up with such force that it comes through the nostrils as well as the mouth. At the same time the bowel, which has been working more or less regularly, becomes constipated and the weight, which has been rising satisfactorily, ceases to increase and very soon begins to fall. An examination of the abdomen shows peristalsis of the stomach and characteristic thickening of the pylorus is felt. The baby is usually breast-fed, a male child, and the first-born, and usually comes from distinctly nervous parents. He has usually been born at full time after a normal pregnancy and normal labour. It is not at all uncommon for one or two members of the same family to have been affected. Thomson, Tribble and Roche cite cases where one, two and three members of a family were affected. The majority of cases begin within the second week to the sixth week, and the history elicited from the mother is, that a previously healthy baby begins to vomit persistently and /

and forcibly, this being associated with marked constipation, the passage of little urine, and a progressive loss in weight. The vomiting is characteristic. It is projectile and explosive and the mother says the milk shoots out on to the floor, from the mouth and nose, a distance of several feet. It differs from the usual vomiting seen in young babies in that it occurs soon after nursing, or while the child is still at the breast, irrespective of whether milk or ordinary water has just been given. Apart from the vomiting there have usually been very few ordinary signs of dyspepsia. The tongue is clean, there are no sour eructations, no abdominal distension and usually no bile in the vomit, no diarrhoea, no pyrexia, no impairment of appetite as the child eagerly looks for a further feed after the stomach has been emptied. Changes of food, unfortunately, cause a temporary relief in the vomiting, but, in a day or two, the vomiting returns as severe as ever and so another food is recommended, which invariably delays the opportunity for early operative treatment. This relief of symptoms is probably due to relief of spasm. The periods of remission of symptoms may be longer than a few days and these cloak the true nature of the case. As the vomiting continues there is generally a steady loss of weight, and the amount of this, along with that of the faecal matter passed, are /

are of great importance because they indicate the degree to which the narrowed pylorus is preventing the passage of food into the bowel, and consequently the degree of urgency of the case. Curiously, the child does not appear ill, though he may be wasted, as, in the early stages, the elasticity of the tissues remains unimpaired. A characteristic feature, however, is the pallor which is nearly always present and, in the later stages of the disease, a drowsiness with shallow breathing, and long periods of apnoea due to accompanying alkalosis.

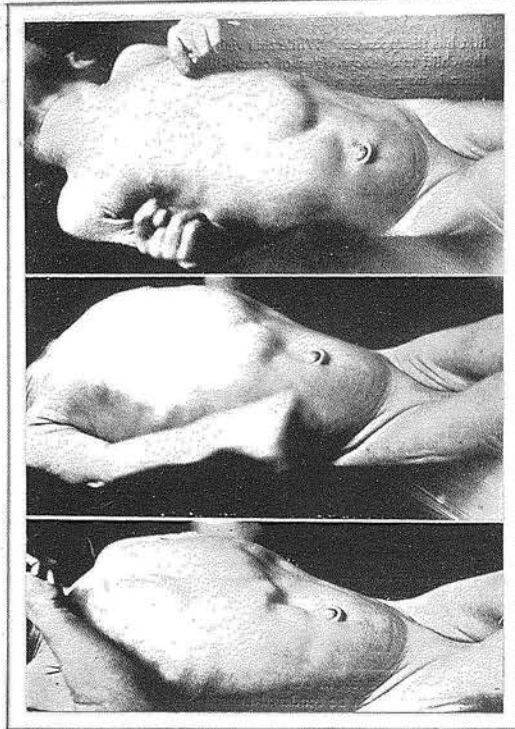
The above symptoms in a child under four months should always lead to the suspicion of pyloric stenosis and demand a search for the pathognomonic features of the disease.

1. Marked, visible gastric peristalsis.
2. Palpable tumour.

Gastric peristalsis is present in every case of pyloric stenosis and is best seen in the gastric region as inspected after the ingestion of some water or milk while the child is quiet.

(Cf. Fig. XI).

Fig. XI. (After Thomson and Findlay).



Deep irregular recurring waves about the size of a golf ball are seen passing from left to right. The size of the wave has no bearing on the degree of obstruction, however. The waves begin at the left costal arch and cross the epigastrium and disappear in the right hyperchondrium. If the stomach is percussed and auscultated it is found to be much dilated. If the child is not comfortable, and wriggles about, contractions of the muscles of the abdominal wall will obscure the peristaltic waves. The abdomen should be kept warm and gentle friction, or tapping over the stomach, or dropping a few drops of ether, often help to set up peristalsis. The peristaltic waves must be marked in order that they may /

may be diagnostic. In any child the stomach contents should stand out when it is full and in any much emaciated child slight peristaltic waves may be seen. Peristaltic waves not only indicate pyloric stenosis but a hypertrophy of the stomach wall as well.

To examine for the pyloric tumour the baby is best lying on the lap of the mother, or nurse, in a warm room. The hand is then placed over the abdomen and gradually explores the pyloric region. The tumour is about the size of the terminal phalanx of the finger. It is situated above, and to the right of, the umbilicus, and varies in consistence depending on muscular contraction. At times it may lie high up under the free border of the liver and sometimes great difficulty is experienced in detecting the tumour. It is wise to search for the pylorus when the stomach is full, if not successful, to re-examine when the stomach is empty. The value of palpation of the tumour as a diagnostic sign has been variously estimated by different writers. Realising that palpation of these tumours is a most important sign we failed in one or two of the cases submitted in this paper owing to the pylorus being tucked under the liver and necessitating elevation of the left lobe with rotation to expose /

expose the pylorus. We felt that other symptoms were so conclusive that we did not delay operation in the hope of a suitable opportunity arising for eliciting the presence of the tumour by palpation.

The secondary symptoms, constipation, loss of weight, scanty urine, are simply results of vomiting. Loss of weight is a constant symptom, and its rapidity is the best guide to the seriousness of the case. In mild cases vomiting may occur only once or twice a day, and, at times, there may be none for a few days. The weight may be stationary or there may be a slight gain. Gradually, in the course of a few months, the vomiting ceases, peristaltic waves disappear and the child, to all appearances, recovers. In the severe cases vomiting is frequent, loss of weight extremely rapid, and the child, if untreated, usually dies of inanition from two to six weeks from the onset of symptoms. Between these two extremes, cases of every degree of severity are seen. The infant is likely to recover if the disease has lasted more than four months. There is no way of predicting when improvement will occur, nor is it possible to predict the degree of severity during the early stages of the illness as an apparently mild case may suddenly become severe.

The association of spasms in other sphincters is not /

not uncommon. Cardiospasm is often recorded in association with pyloro-spasm. In one of our cases cardio-spasm, as well as pyloro-spasm, was evidenced on an X-ray plate after a bismuth meal.

DIAGNOSIS.

It can be confidently stated that no disease of infancy can be more frequently and correctly diagnosed so long as one is alive to the possibility of its presence. If this condition is kept in mind the diagnosis may be made formally in almost every instance, and is not a matter of opinion but is demonstrable. No laboratory tests, or other procedures requiring the services of specialists, are necessary. The general practitioner is as well able to diagnose a case as a paediatrist, if the fundamental symptoms and signs of the condition are familiar. The history, if obtained from an intelligent mother, is valuable and often typical, but such a history must be very carefully and fully taken. The characteristic projectile vomiting is frequently observed during examination. A palpable pyloric tumour and gastric peristaltic waves in combination occur in no other condition. The presence of alkalosis, evidenced /

evidenced clinically by slow, shallow breathing, or a chemical examination of the blood by its increased CO_2 content, should always make one suspect pyloric stenosis, as it is found in practically no other infantile disease.

Although diminution of the chlorides in the urine is common in pyloric stenosis it may occur in vomiting from whatever cause and hence is of no diagnostic value. A normal chlorine content of the urine, on the other hand, is distinctly against the presence of pyloric stenosis.

The question of forming a diagnosis by X-ray examination is one that is frequently discussed.

Fig. XII. This shows evidence of pyloric stenosis in a male infant, 5 weeks old. Plate taken five minutes after opaque meal.

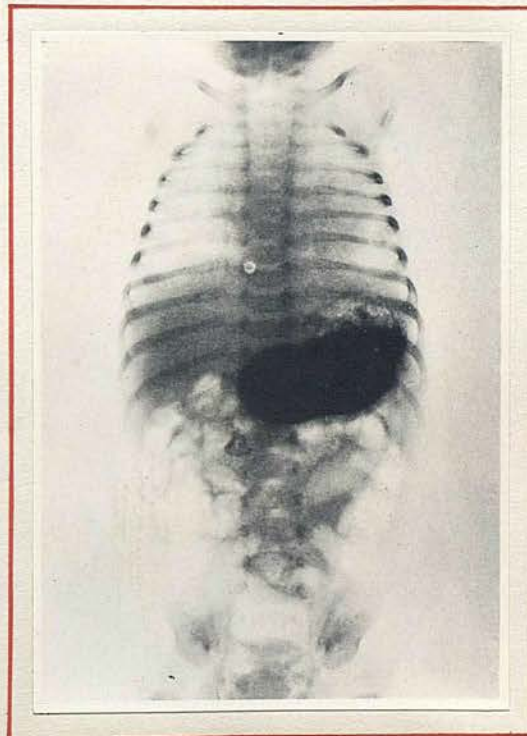


Fig. XIII. The same patient as in Fig. XII. X-ray taken four hours after opaque meal. There is no dilatation of the stomach but considerable retention.



In our opinion, however, the diagnosis of pyloric stenosis may be made without the aid of X-ray, especially as a barium meal is not without its dangers.

The cases most difficult to distinguish from pyloric stenosis are those of pyloric spasm. The patient is usually a female child; much more evidence of pain, vomits after each feed, the whole contents of the stomach are returned at once or else it recurs at short intervals until the stomach is empty. There is no tendency for food to accumulate in /

in the stomach, no signs of gastric dilatation or hypertrophy; there is never any characteristic peristalsis to be seen, motions and urine are scanty, sometimes slight diarrhoea occurs. In cases where history is atypical and no tumour is palpated, an opaque meal is used to confirm the absence of gastric retention, and in those infants who vomit from birth the early use of X-ray is imperative. In such cases there is a strong possibility of congenital atresia of the duodenum, which the X-ray will reveal.

CHOICE OF TREATMENT.

In all cases, unless perhaps those seen within a few days after the onset, medical measures should be adopted in the first instance. Even those cases which are in a sufficiently good state for immediate operation improve if they are treated medically for twenty-four or forty-eight hours. The treatment consists in lavage of the stomach, administration of a suitable diet, and replacement of loss of fluid which may have taken place. The medical measures which have been carried out by us are the administration of atropine, gastric lavage, and thick cereal feeding if breast milk is unobtainable. Whatever the results of medical treatment are there can be no doubt /

doubt that a large number of these cases require to be in Hospital for a period of weeks or months under careful medical supervision and expert nursing, and the dangers of protracted hospital care in individuals are well-known. Medical treatment demands the weaning of the baby which, in itself, is not a bad feature, if the breast milk can be encouraged to continue. It is advisable that the breast milk be extracted and given in measured quantities by the bottle, rather than that the child should be permitted to drink direct from the breast. Most observers agree that the mortality is greater in artificially fed babies than in breast milk babies, and in this series of cases, towards the end of the treatment, on many occasions breast milk was unobtainable, and we had to resort to artificial food mixtures after the first week following the operation. We were fortunate in being able to secure a fairly liberal supply of breast milk from our maternity hospital which adjoins the Children's Hospital. At times, however, this was not obtainable and artificial feeding had to be resorted to.

As soon as the diagnosis of pyloric stenosis is established, and the infant is operated on, it is usual for the child to be home and on the breast again /



again within a week. Operative mortality is largely due to a delay in diagnosis, or in choice of surgical treatment, or imperfect preparation for operation. In view of the uncertainties which beset medical treatment there appears to be little justification for any delay which turns a good operative risk into a bad one. In the case of an infant under two months of age, in whom the diagnosis of pyloric stenosis is made, the indications, in our opinion, are clear for prompt operation.

Babies seen for the first time in the third or fourth month afford the real problem in the choice of treatment. When weight loss is persistent the indication is clear for operation, up to the end of the fourth month. Medical treatment should be persisted in if the weight is gradually increasing and if the infant is only slightly under weight, unless such treatment involves weaning the baby, in which case surgery is to be preferred. In severe cases seen in the latter part of the fourth month the outlook is not good for any form of treatment.

PRE-OPERATIVE TREATMENT.

Following on the work done by Maizels, McArthur and Payne, and Clopton and Hartmann, greater knowledge of blood chemistry has been made possible, and, as a result /

result of this work, considerable attention is now paid to pre-operative care.

Most of the cases are brought to the hospital markedly dehydrated, and at times showing alkalosis.

To overcome these conditions abundant subcutaneous injections of Ringer's solution are necessary. If athrepsia is present, glucose solution intravenously, or whole blood transfusion, is indicated.

We regard the preparation of the patient for operation as one of the most important factors in the handling of these cases.

Usually twenty-four hours or more is given to the replacing of fluid loss and the counteracting of starvation symptoms before we dare operate. Hence the major part of the battle is fought before we enter the operating room.

In general the factors which tend to increase the operation risk are:-

(1) Disturbance of the acid base equilibrium of the body.

(2) Anhydraemia.

(3) Marked asthenia due principally to malnutrition and anaemia.

(4) Presence of infection.

Marked vomiting, when due to pyloric stenosis, causes loss in the vomitus of hydrochloric acid usually in large /

large amount and of the base chloride in smaller amount.

The loss from the body of the chloride ion is almost invariably compensated for, in a large part, by retention of the bicarbonate ion in the blood and tissue fluids, leading to alkalosis of varying severity. Such a shift toward the alkaline side is just as serious as a commensurate shift to the acid side (acidosis) if not more so.

Death may occur promptly as a result of collapse, cessation of respiration, or generalised convulsions with laryngeal spasm.

Aside from loss of acid by vomiting, alkalosis may be increased by any measure which would tend to cause exaggerated breathing.

Such instances are commonly seen as a result of crying, because of hunger, pain or manipulation. It is important then, since all those factors may be present before, during, or immediately after operation, to restore if possible the acid base balance of the body to its normal equilibrium before operation.

The diagnosis of alkalosis can be made both clinically and chemically. In the first place, alkalosis of some degree almost invariably accompanies marked vomiting due to some type of obstruction of the gastro intestinal tract. It is therefore always expected

expected in cases of pyloric stenosis especially if breathing is shallow, depressed, and irregular with frequent apnoeic pauses.

Further evidence of alkalosis might be noted in the appearance of general hypertoxicity and such evidences of tetany or carpopedal spasm, positive chtvostek sign or generalised convulsions.

The urine characteristically is free from chloride (when acidified gives little or no white precipitate after addition of A_2NO_3) but also contains so little base bicarbonate (BHCO_3) that it is distinctly acid (pH_{5-6}) in reaction. This latter point is of importance.

Ordinarily, alkaline urine will rule out the presence of acidosis of any type except that associated with nephritis, but acid urine not only does not rule out alkalosis, when of the type associated with vomiting, but its presence actually lends support to the diagnosis. Certain diagnosis of alkalosis, however, can be made by chemical examination of the blood.

Increased pH (alkalinity) and CO_2 content (base bicarbonate) will be associated with diminished base chloride.

Not only does the disturbance of the acid base balance tend to produce tetany and its consequences but it also contributes to anhydraemia.

In /

In addition to the fluid loss of food intake there occurs the loss of water bound to HCl. or BCl.

These two factors, i.e. disturbance of the acid base balance and anhydraemia, are therefore closely associated and the treatment of both is carried out at the same time.

The treatment in general consists of the administration of water and salt in the form of Ringers solution. As a rule, Ringers solution is administered subcutaneously two or three times daily until the chloride content of the blood remains at approximately the normal level. When this occurs the bicarbonate content of the blood will have dropped to approximately the normal level because of the excretion of bicarbonate into the urine which now renders the urine highly alkaline. At the same time anhydraemia disappears.

In a very few cases, showing very marked chloride reduction, it may be necessary to give a more concentrated salt solution such as 3% sodium-chloride subcutaneously but this rarely has to be done.

If at any time acute manifestations of alkalosis are seen, such as tetany or marked depression of the respirations, immediate measures directed toward relief of these acute symptoms will have to be instituted. They are:-

Breathing /

Breathing of 30% CO_2 in oxygen. This is a quick and effective means of stimulating respiration, of providing sufficient oxygen to saturate the haemoglobin and to increase the free carbonic acid of the blood, so that the rates of $\text{NaHCO}_3 : \text{H}_2\text{CO}_3$ becomes more nearly normal due to increase in the denominator.

When this happens the actual signs of tetany are relieved.

In addition a 5% calcium chloride solution in an amount equivalent to $\frac{1}{2}$ c.c. per kilogram body weight is given intravenously. Calcium given in this way is also immediately effective in relieving tetany.

A drug which acts in a similar manner and which is sometimes used is a 10% anhydrous magnesium sulphate given in 2 doses each of 1 c.c. per kilogram of body weight 15 or 20 minutes apart.

Magnesium is just as effective as calcium in relieving tetany, but has one distinct disadvantage in that it tends to depress the respirations which, because of the presence of alkalosis, are frequently already depressed. If, however, depression of respiration follows the administration of magnesium sulphate, calcium chloride may be given as an antidote to magnesium sulphate for the depressant effect.

Occasionally an infant is so weak, because of starvation and anaemia, that he would be a very poor operative /

operative risk. In such instances frequent intravenous injections of from 10 to 20% glucose are given, very slowly, so as to provoke as little diuresis as possible. Fluid and some immediate food in this way are given.

Blood transfusions are also resorted to and, in addition, an attempt is made to give the infant the usual thick cereal feedings, preceded by atropine in physiological doses.

OPERATIVE TREATMENT.

Preparation for operation. Before operation the limbs are bandaged in cotton wool, and a special pad of cotton wool applied to the chest, sufficiently small not to interfere with the operative area but yet able to retain the necessary warmth over the vital centres. The theatre and table are specially warmed and measures are taken for the prevention of shock. About half an hour before the baby is moved to the theatre the stomach is washed out with normal saline. This is done for various reasons:-

(1) the ease with which it is possible to handle an empty stomach.

(2) Should a puncture be made in the mucous membrane the risk of infection is less.

(3) /

(3) On the re-establishment of the pyloric canal, stagnant food and mucus do not pass into the easily irritated intestine.

(4) Should regurgitation occur during the operation the danger of respiratory obstruction is lessened. It is advisable to wash out the stomach with a number 14 catheter to clear it of all plugs of mucus.

(5) Normal saline is used instead of bicarbonate of soda which tends to induce alkalosis.

Anaesthesia. Anaesthetic is a matter of great importance, gas and oxygen, with occasional resort to short periods of ether vapour, has been the anaesthetic of choice. The infant must be sufficiently anaesthetised to prevent shock and also to avoid straining and protrusion of the omentum or viscera. If this occurs the shock is increased, the operation delayed, and difficulty is experienced in replacing the protrusion and stitching up the peritoneal opening.

We have never varied in our method of anaesthesia, though we are fully aware many surgeons prefer local anaesthesia. We have felt that, no matter how carefully a local anaesthetic is administered, there is always a slight element of tissue destruction present from the traumatising effect of the injected fluid /

fluid and this would tend to lessen primary healing. As our results have been so satisfactory with general anaesthesia we have felt it wiser to persist in the method we are accustomed to.

Operative Technique. An incision is made $1\frac{1}{2}$ to 2 inches long, slightly to right of the midline through the rectus muscle. It is placed fairly high, just below the level of the ensiform cartilage. On opening the peritoneum the lobe of the liver is seen. This is rotated upwards to expose the stomach and at the end of the operation the liver falls back to its natural position, fills the wound, and so helps to prevent protrusion of abdominal contents. As soon as the abdominal cavity is opened the edges of the peritoneum are secured before they retract. This avoids delay when the time for closure comes. When the lower edge of the liver has been hooked upwards the stomach appears in the wound. The diagnosis having been confirmed by feeling the pyloric tumour with the finger introduced into the abdomen, the tumour is hung up by the examining finger, and brought into the wound, and the stomach is pushed upwards into the abdomen. The pylorus being delivered the right end of the incision is determined by the always fairly visible junction of the /

the whiteish, opaque, solid pylorus with the bluish, translucent, flaccid duodenum. The cut should begin 1 mm. or 2 mm. short of this and extend well into the pyloric antrum, curving upwards in its left third to follow the bloodless line between the superficial blood vessels. Most illustrations give the impression of too short an incision and, as there is no definite anatomical limitation in this direction, there is no reason for risking possible blockage here by curtailing the cut. The mucosa is then exposed by blunt dissection, but exposure is far from being enough. The aim of the operation is not so much to allow the passage of food through the original pyloric canal as to form a totally new one alongside it by the bulging out of the uncovered mucosa. To do this, at least half an inch of it should be exposed in the middle of the incision and as near as possible to either end. By far the easiest and safest way to do this is by stretching the edges of the incision in the muscle apart so that they tear away from the intact mucosa beneath owing to the looseness of the subjacent areolar tissue. This is perfectly easy and safe to do over the pylorus and on to the stomach, but it is a different matter at the duodenal end. Here the zone of adhesion is in danger as, although the muscle will separate from the mucosa as easily in /

in the duodenum as in the stomach, there lies between these two regions this ring in which serosa, muscle, and mucosa are all bound together and will strip together or not at all. If it were not for this it would be possible to divulse the muscle just as easily at this end of the wound as at the other and to leave the mucosa to take care of itself. In avoiding this dangerous region there are two aids. The first is to remember that it is marked by the vessel described and lies superficially so that, while it is risky to extend the wound near the surface, it is comparatively safe to stretch the deep muscle fibres till they part. The second aid is the recognition of the annular constriction which is announced by the mucous membrane ceasing suddenly to bulge when it is released and dipping down into the choke described.

When the muscle has been completely divided the edges of the incision retract and the redundant mucous membrane bulges freely into the gap. Haemorrhage is controlled by the injection of one of the serum preparations, usually haemostatic serum, into the hypertrophied muscle. If this is not sufficient the vessels may be under-run and ligatured. The patency /

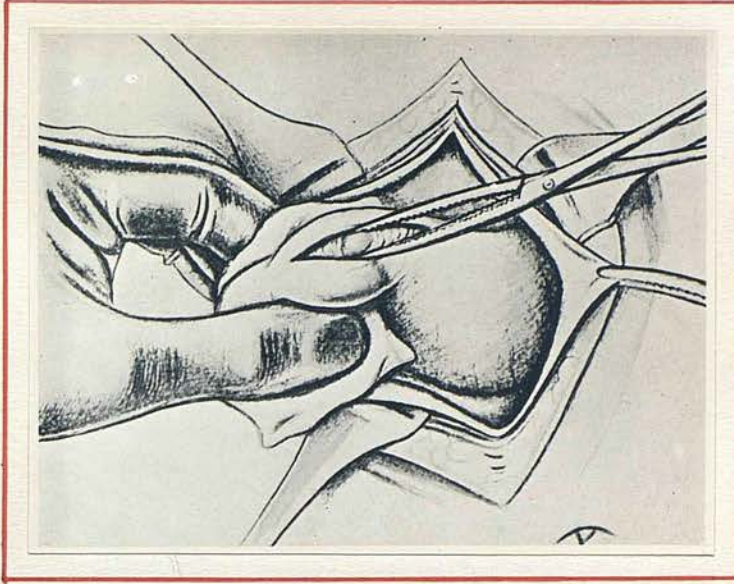
patency of the canal may be tested by gently squeezing the stomach and this precaution also serves to demonstrate the absence of microscopic perforation of the mucous membrane. The pylorus is now returned to the abdomen and the liver allowed to fall into place. The closure of the wound is begun at the lower end, since it is here that protrusion of the abdominal contents is most likely to occur. A continuous suture closes the peritoneum and muscle in one layer. Three or four deep stitches through the skin and muscle external to the peritoneum are now inserted. A continuous suture of catgut closes the opening in the rectus sheath. The deep sutures are tied over rubber bands and the intervening portion of skin approximated with michel clips.

Fig. XIV. (After J. V. Donovan).



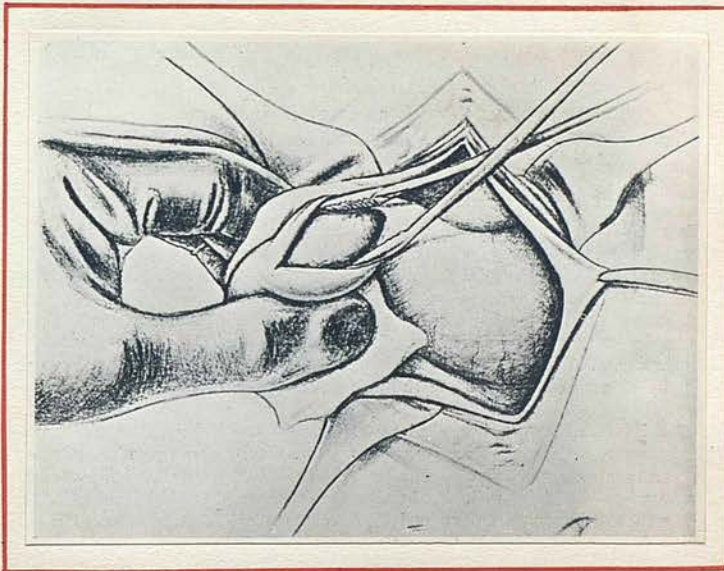
This diagram shows an abnormal pylorus. An incision is being made into the hypertrophied muscular tissue down to the mucosa.

Fig. XV. (After J. V. Donovan).

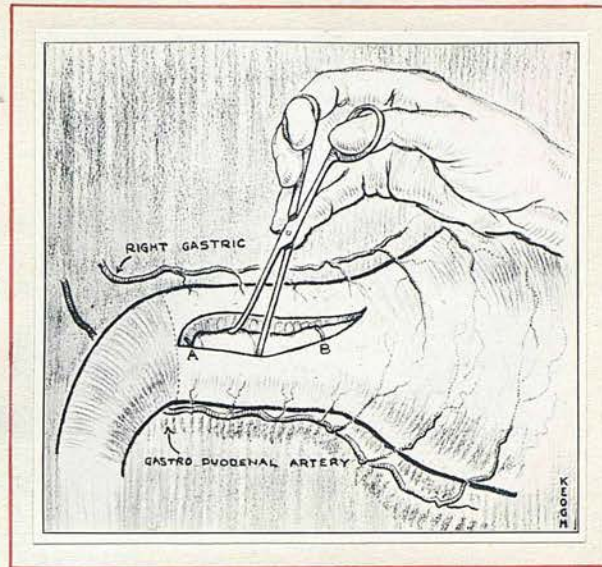


This illustrates the separation of the divided muscular tissue with a pair of sinus forceps. By this method the muscle tears readily and prevents any possibility of opening into the mucosa.

Fig. XVI. (After J. V. Donovan).



Illustrates the completed stage of the operation. The muscle has been divided up to the duodenal margin and well into the pyloric antrum. The mucosa now bulges through the incision and is usually level with, or projecting beyond the cut margins of the hypertrophied muscle.

Fig. XVII..

This is an illustration from Dennis Browne's article on the Surgical Anatomy of the Pylorus when affected by congenital hypertrophy.

- a. Represents the minute vessel seen at the duodenal end of the hypertrophied area.
- b. The small vessel seen at the gastric end of the hypertrophied area.

AFTER-CARE AND FEEDING.

We are in entire agreement with Hughes who urges the closest co-operation between the surgeon and the paediatrist in the after care of these cases. The bowel having been in disuse for some time, is unable to cope with indiscretions in the diet; for this reason every effort possible must be made to obtain human milk for at least the first week after operation. Although /

Although in about half our cases the Mother still had milk, we were able in all the others to obtain reliable human milk from some other source. In consequence there were no diarrhoea complications or vomiting to deal with. It is impossible to over-stress the value of human milk during the first week.

If human milk is definitely unobtainable, a mixture of buttermilk two parts, water one part, and a 5% to 10% addition of Dextri-Maltose is given. After a week the buttermilk is replaced by an acidified full milk and the increases in feeding carried on in accordance with the progress of the infant.

Immediately on return from the theatre 2 ozs. of Ringer's Solution with 5% Glucose is given very slowly per rectum, and repeated six-hourly during the first twenty-four hours, provided the infant is retaining the injections. One hour after operation a teaspoon of water with 5% Glucose is given by pipette, and followed, an hour later, by a teaspoonful of human milk given in the same way. Drachm doses of milk are given hourly for the next ten hours with Glucose Drinks in between. During the next twelve hours 2 drachm doses of milk are given hourly if the child is awake, while on the next day two-hourly feeds of half an ounce are given.

Then, if the child is progressing favourably, the /

the Mother is allowed to lean over the cot and allow the infant to suckle for five minutes. We find the early suckling greatly assists in maintaining the supply of milk in the breasts. Feeds are gradually increased, according to the condition of the case, until the fourth or fifth day - 2 ozs. feeds are being taken two-hourly or $2\frac{1}{2}$ ozs. three-hourly.

If the progress is normal and the Mother has sufficient milk herself, the infant is sent home on the fifth day and returns on the eighth to tenth day for the removal of the stitches.

Cases which will eventually be fed on cow's milk are given a mixture of cow's milk mixed with human milk from the fifth day, and by the ninth day only a cow's milk mixture is given and the case discharged by the fourteenth day.

COMPLICATIONS.

1. Shock. This must be most carefully attended to pre-operatively as these children are usually in a very low state of health and able to withstand little interference. The rapidity and ease of the operation, however, in a properly prepared case, do not tend to produce any marked degree of shock.

2. Haemorrhage /

2. Haemorrhage. This is unusual but is cited as a definite complication. If it should be at all troublesome it is best controlled by (1) injection of serum; (2) ligatures; (3) the muscle graft; (4) omental graft. It is seldom necessary for more than one or two of these methods to be used before all haemorrhage is completely controlled.

3. Peritonitis. This is a complication which we have not met but which has been described. It might, of course, be due to faulty asepsis but more than likely is the result of a minute perforation of the mucous membrane which has escaped notice at the operation.

4. Diarrhoea. Diarrhoea, accompanied by vomiting, was fortunately not frequent in our series of cases, but may easily be fatal. The two factors responsible for the onset of Diarrhoea are (1) the intestine of such an infant in its short life before the operation has not exercised its full normal functions and (2) since normal quantities of food have not passed out of the stomach it seems reasonable to suppose that the intestinal mucous membrane is more susceptible to irritation than in the case of the normal infant and has acquired very little immunity to the cause of infective diarrhoea.

6. Post-Operative Fever. Another complication which may cause a fatal result is hyper-pyrexia. It is commonly encountered after a Rammstedt operation, the temperature rises sharply to 101° or to 103° and falls again in from 24 to 48 hours. Sometimes, however, the temperature may rise as high as 107° or 108° and cannot be reduced even by vigorous treatment so that death rapidly follows. It is well-known that the heat regulating mechanism of infants is easily disturbed and that this instability is increased in an undeveloped, wasted or exhausted patient. One would also expect that the susceptibility to irregular temperature would be in the proportion of the degree of wasting and exhaustion present. A partial explanation offers itself in the lack of body fluids and consequent diminished perspiration.

In the first half of this group of cases a post-operative rise of temperature occurred. This was not excessive, soon fell, and had no unfavourable result, but Ramsay states a case in which the temperature rose to 108° and the child died of convulsions /

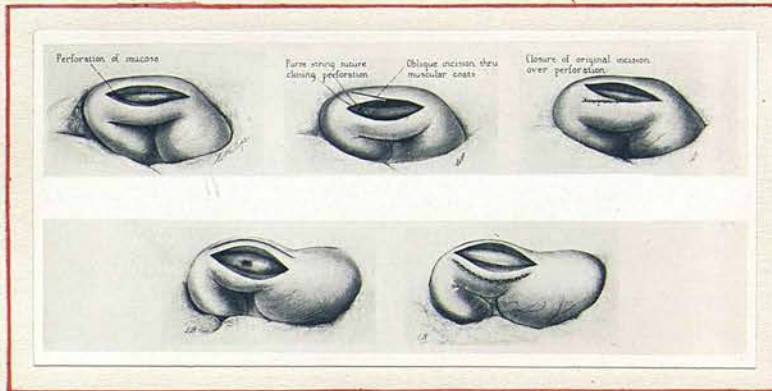
convulsions in 26 hours. He considers that these wasted infants are so susceptible to irregular fever, the same factor being responsible for the onset of pyrexia and diarrhoea. The intestine is unused to food and its resistance to extraneous substances such as food, and its absorptive powers are not normal. Thus it seems possible that the sudden influx of food in an unaccustomed quantity may, by local irritation and abnormal absorption initiate a rise of temperature which can easily become uncontrollable on account of the unsatisfactory general state of the patient. An interesting case is cited by him of an infant which was operated on for pyloric stenosis by the usual Rammstedt operation. There was no post-operative rise of temperature and the symptoms were not relieved, vomiting continuing, and the child going downhill. On the fifth day the abdomen was reopened and obstruction was found to be unrelieved. A more satisfactory incision was made in the pylorus and the abdomen closed. The symptoms ceased and the child recovered. An interesting point, however, is that, after the second operation, a typical rise of temperature occurred. This case suggests that the passage of food into the intestine has some influence on the post-operative temperatures. With the /

the newer methods of pre-operative treatment this tendency to hyperpyrexia is very much lessened as abundance of fluid has been given prior to operation and the child is in a better state to withstand surgical interference.

Diarrhoea and high fever are the most frequent causes of death after operation, and it appears to be due primarily to the atrophied condition of the intestine consequent upon partial or complete stoppage. The longer such starvation is continued the greater the risk and this is one of the main reasons for urging that no time be lost before undertaking surgical treatment in those cases in which medical methods do not show early signs of success.

7. Otitis Media must be associated with pyloric stenosis. Presumably the high incidence of otitis media in these cases results from the vomiting and forcible injection of the vomitus into the eustachian tube. Operative risk in such cases will be lessened if the otitis media is recognised and treated as it would be in any other cases.

Fig. XVIII. (After O. F. Lamson.)



This diagram is to illustrate a very satisfactory method of closing a perforation in the mucous membrane which may occur during the performance of a Fredet-Rammstedt operation. The diagram is self-explanatory of the technique recommended to be used.

TABLE OF CASES.

<u>Case</u>	<u>Date</u>	<u>Sex</u>	<u>Hospital or Private</u>	<u>Age at Operation.</u>	<u>Weight at Operation.</u>	<u>Age of Onset of Symptoms.</u>	<u>Progress.</u>	<u>Result.</u>
1. J.W.	15/5/26	Female	Private	7 weeks	7 lbs. 6 oz.	3 weeks	Breast-fed, uneventful.	Cured
2. R.B.	19/10/26	Male	Private	8 weeks	7 lbs. 8 oz.	3½ weeks	(Breast milk, { artificial { milk. Breast-fed.	Cured
3. D.C.	2/8/27	Male	Private	7 weeks	9 lbs. 6 oz.	3 weeks	Breast-fed.	Cured
4. B.E.	16/4/28	Male	Private	8 weeks	8 lbs. 12 oz.	3½ weeks	(Artificial (feeding. Bron- (cho Pneumonia.	Death
5. H.T.	23/10/28	Male	Private	3 weeks	6 lbs. 8 oz.	2½ weeks	Breast-fed.	Cured
6. I.C.	12/11/28	Male	Private	11 weeks	10 lbs. 10 oz.	3½ weeks	Breast-fed	Cured
7. G.C.	13/3/29	Male	Private	6 weeks	6 lbs. 10 oz.	2½ weeks	Breast-fed	Cured

TABLE OF CASES (CONTD).

<u>Case</u>	<u>Date</u>	<u>Sex</u>	<u>Hospital or Private.</u>	<u>Age at Operation.</u>	<u>Weight at Operation</u>	<u>Age of Onset of Symptoms.</u>	<u>Progress.</u>	<u>Result.</u>
8. P.O.	8/4/29	Male	Hospital	12 weeks	6 lbs. 12 oz.	3 weeks	Collapse	Death
9. S.O.	3/1/30	Male	Private	3 weeks	6 lbs. 8 oz.	2½ weeks	Breast-fed	Cured
10. B.C.	16/3/31	Male	Private	7 weeks	7 lbs. 2 oz.	3 weeks	Vomited after Operation	Cured
11. T.L.	16/2/32	Male	Private	8 weeks	8 lbs. 2 oz.	3 weeks	Breast-fed	Cured
12. P.L.	24/3/34	Female	Private	4 weeks	6 lbs. 6 oz.	2½ weeks	Artificial Feeding	Cured
13. C.S.	15/2/34	Male	Private	7 weeks	7 lbs. 7 oz.	3 weeks	(Breast-fed, (Artificial (fed.	Cured
14. J.F.	16/5/34	Male	Private	7 weeks	7 lbs.	3½ weeks	(Breast-fed, (Alkalosis, (Tetany.	Cured

S U M M A R Y.PATHOLOGY.

We feel we have shown, by histological sections of the normal pylorus and of the pylorus affected by congenital hypertrophy, that the pyloric sphincter itself is not directly affected by the hypertrophy.

The prepyloric portion of the pyloric canal undergoes marked hypertrophy of its circular muscle, and we believe an outgrowth of this circular muscle, forcing its way into the duodenum, compresses the pyloric sphincter, which is only slightly enlarged and is seen to be at the apex of the fornix of the portio vaginalis. The points which occur to us are such questions as, whether the pyloric sphincter and the prepyloric portion of the pylorus have a separate nerve supply.

Is this under the control of the sympathetic-parasympathetic nervous system? We know in Hirschsprung's Disease division of the sympathetic fibres leads to remarkable recoveries.

More recent work on cardiospasm shows very encouraging results by removing the sympathetic fibres round the left gastric artery and its oesophageal branch.

In no other condition, however, is there such marked hypertrophy of muscle at the sight of obstruction /

tion, and it seems doubtful even if the course of the sympathetic fibres to the pylorus was definitely established, that attempts would be made to divide these nerves in the hope of a satisfactory result. The treatment, by division of this hypertrophied muscle, is so satisfactory in this condition, that little in the way of methods of treatment is being sought after, but more in the nature of methods of prevention. The causation, however, still remains definitely obscure.

CLINICAL.

This thesis is based on fourteen cases operated on over a period of nine years. Two of the cases were delivered into this world by the author and these were the only cases operated on at an early date. The average age of these infants when submitted to surgical treatment was $6\frac{1}{2}$ weeks and the average weight was 7 lbs. 8 ozs. In all cases a Fredet-Rammstedt operation was performed.

The majority of these infants were discharged from hospital within seven to nine days.

Two of these babies died, one from broncho pneumonia 14 days after the operation, and the other immediately /

immediately after operation owing to the parents refusing operation when the child was four weeks old, and returning two months later with the marasmic, dying infant, pleading for something to be done. The prognosis in this case was extremely bad as the child was practically moribund and at that time our knowledge of pre-operative treatment was very much less than it is now.

The work of Maizels & McArthur, who stress the value of pre-operative treatment and the recognition of alkalosis so common in neglected pyloric stenosis cases has, we feel, done much to make the course of these little patients much easier after operation and has resulted in a greatly lowered mortality.

Another important factor is the advantages of the private room in hospital and the private nurse.

Most of these cases were operated on in the Transvaal Memorial Hospital for Children.

All were in private rooms and many had their own private experienced children's nurses.

Congenital pyloric stenosis has always been looked upon, and we believe rightly, as a medical disease.

The operation is only an incident, albeit an important one, in the malady.

The /

The gradual onset, the change from occasional regurgitation to the characteristic vomiting, the presence of gastric peristalsis and the routine palpation of the abdomen for a tumour must be the duty of the physician.

Similarly, after the operation, the details of feeding are of great importance to secure a successful result and entirely beyond the province of the surgeon.

The surgeon may study his operation figures and decide that his failures occur in wasted infants who have been weaned from one patent food to another. That his successes occur in the breast fed children of nearly normal weight and in his private cases which are nearly always diagnosed and operated on at an early age. It is reasonable for him to ask that his aid should be invoked before the patient is in extremis.

We agree most heartily with Wallace & Wevill who show, in a report of 145 pyloric stenosis cases operated on in the Royal Hospital for Children, Edinburgh, that if the mortality rate which in this series was in the region of 20% is to be reduced to a reasonable figure, it is essential that the diagnosis should be made at a much earlier date.

There /

There appears in nearly all cases to be a period of three to four weeks after the commencement of symptoms, during which time these cases are treated as digestive disorders and the feeding is changed every few days in the vain hope that the vomiting will cease. Eventually, when the child is more or less in extremis, it is sent to hospital in a state of health which makes any form of treatment a dangerous procedure.

Pyloric Stenosis should be considered as a possibility in every case of forceful vomiting in infancy and it should be recognised that the longer the delay in sending the child for treatment the less are its chances of survival.

It should be considered an abdominal emergency of infants and one feels confident, if the condition were diagnosed within one week after the commencement of symptoms and sent for operation without delay, the mortality rate would be greatly reduced.

How can this be attained? We feel it is only from the family doctor, more keenly alive to this somewhat rare disease and more aware of the possibilities of surgery, that further improvement must come.

In /

In conclusion, I desire to express my thanks to Professor D. P. D. Wilkie for allowing me the privilege of working in the Surgical Research Department of the University of Edinburgh; and to Miss Agnes MacGregor, Pathologist to the Royal Sick Children's Hospital, Edinburgh, for her helpful advice on the study of the Pathology. I also wish to thank Mr F. W. Pettigrew and his technical staff for their skilful help in preparing the specimens, photographs, and micro-photographs; and Mr Clifford Shepley for his excellent drawings.

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